Echocardiographic Features of Right Heart Failure in Infancy and Childhood

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In infants and children, right heart failure is most frequently a consequence of increased afterload (pulmonary hypertension). However, it is also observed as a sequela of congenital cardiovascular surgery. The purpose of this report is to present the causes of right heart dysfunction in children and to define the echo-Doppler methods used in the evaluation of right heart failure. (ECHOCARDIOGRAPHY, Volume 15, No. 8, Part 1, November 1998)

right ventricular failure, pulmonary artery failure, congenital heart disease

For the purposes of discussion, the etiology of right heart failure is divided into primary and secondary causes. Primary right heart failure results from either a congenital malformation of the heart or peripheral vascular system. In contrast, right heart failure is considered secondary when it occurs as a response to airway disease. The physiology of right ventricular failure is similar in both circumstances. Reduction in right heart function results from increased preload, afterload, or both.

The pyramidal shape of the ventricle makes it less able to accommodate physiological stresses, and it initially dilates and then hypertrophies. As failure progresses, changes in right ventricular ejection (systolic function) and filling (diastolic function) become evident. From a practical standpoint, the evaluation of right ventricular function by echo-Doppler in children is not different from that in adults.

Systolic Function

Initial studies used right ventricular systolic time intervals[1,2] to assess right ventricular systolic pressure and function. These ejection phase indices, however, were affected not only by changes in systolic function but also by changes in preload and afterload. Subsequently, estimations of right ventricular volumes and ejection fraction using cross-sectional imaging became popular (Figs. 1 and 2).[3-5] Finally, Doppler techniques were used in estimating right ventricular stroke volume and output.[6-8] Either tricuspid inflow velocity or pulmonary outflow velocity may be used for the estimation of stroke volume.

Stroke volume = mean inflow velocity
× tricuspid valve area × flow time

or

Stroke volume
= mean pulmonary flow velocity
× pulmonary valve area × flow time

where valve area is calculated from the annular diameter, assuming a circular orifice.

Diastolic Function

Reasonable estimations of diastolic function are made with Doppler echocardiography. Right ventricular diastolic filling velocities provide these insights into diastolic function. Normal

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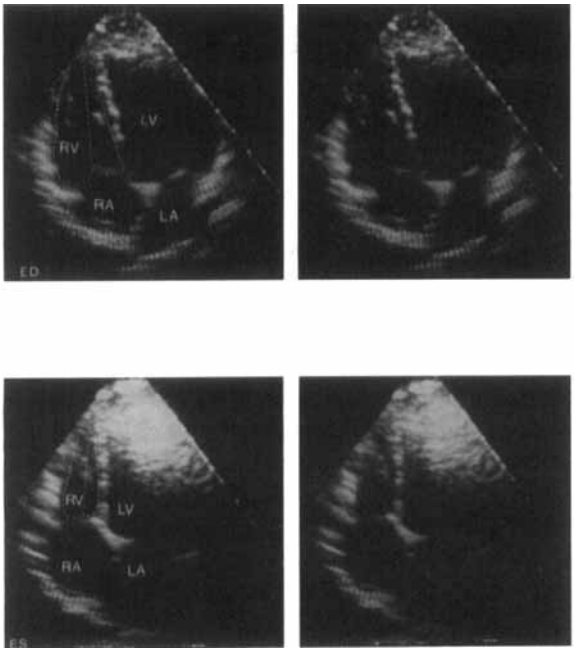

Figure 1. *Apical four-chamber view with end-systolic (ES) and end-diastolic (ED) frames outlined demonstrating the method of planimetry used for calculation of right ventricular (RV) volumes and ejection fraction. LA = left atrium; LV = left ventricle; RA = right atrium. Reprinted with permission from Hiraishi et al.*[4]

tricuspid inflow velocities have been described for fetuses,[9] neonates,[10] and children[11] (Table I). Changes in the peak early inflow velocity and E/A ratio, as well as isovolumic contraction time, may be observed with pulmonary hypertension or right ventricular diastolic dysfunction with normal pulmonary pressures.[12]

Pulmonary Artery Systolic and Diastolic Pressures

The earliest method of detecting pulmonary hypertension involved the use of right ventricular systolic time intervals measured from the pulmonary valve echogram.[1] As pulmonary vascular resistance falls in the newborn, the ratio of the right ventricular pre-ejection period (RVPEP) to the right ventricular ejection time (RVET) decreases due to an increase in ejection time.[13] The normal newborn change in RVPEP/RVET must be accounted for when using this method for the estimation of elevated pulmonary pressures. Recently, color Doppler-directed continuous-wave Doppler was used to predict pulmonary artery systolic and diastolic pressures. With the use of a modified Bernoulli equation ($\Delta P = 4V^2$) and velocity of the tricuspid regurgitant jet or a pulmonary regurgitant jet, the pulmonary artery systolic and diastolic pressures can be calculated.

Pul. systolic pressure

$$= 4(\text{TR velocity})^2 + 4 \text{ mmHg}$$

Pul. diastolic pressure

$$= 4(\text{PR velocity})^2 + 4 \text{ mmHg}$$

where 4 mmHg is normal right atrial mean pressure and right ventricular end-diastolic pressure,[14] Pul. is pulmonary, TR is tricuspid regurgitation, and PR is pulmonary regurgitation.

An alternative method for estimating pulmonary artery systolic pressure uses the maximum velocity of flow across a patent ductus arteriosus.

Pul. systolic pressure

= cuff or UA systolic pressure

$- 4(\text{PDA velocity})^2$

where UA is umbilical artery, and PDA Vel is patent ductus arteriosus maximal velocity.

It is traditional to express pulmonary pressures as a percentage of systemic arterial pressure. Numerous studies have shown the usefulness of Doppler measurements of right ven-

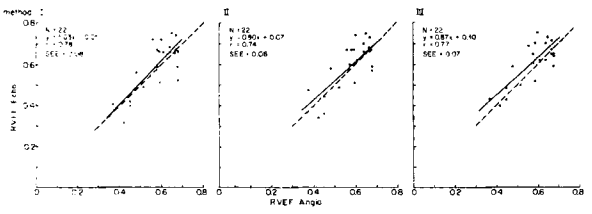

Figure 2. *Relationship observed between echo-calculated right ventricular ejection fraction (RVEF) and angiographically calculated ejection fraction. SEE = standard error of the estimate. Reprinted with permission from Hiraishi et al.*[4]

TABLE I

Right Ventricular In Flow Velocities

	Early Gestation Fetus [9] (17–24 weeks)	Late Gestation Fetus [9] (31–36 weeks)	Newborn [10] (24 hr)	Child (11)	
				Insp	Exp
Peak E (cm/sec)	26.3 ± 2.0	35.3 ± 0.9	46.6 ± 10.0	85 ± 11	92 ± 14
Peak A (cm/sec)	40.9 ± 2.8	45.4 ± 1.2	53.0 ± 8.4	52 ± 12	50 ± 13
E/A ratio	0.63[a]	0.78[a]	0.84 ± 0.14	1.72 ± 0.42	2.0 ± 0.64

[a] Extrapolated from peaks A and E. Exp = expiration; INSP = inspiration.

tricular inflow and outflow velocities as well as right ventricular systolic time intervals in estimating elevated pulmonary artery pressure and vascular resistance.[15,16]

Congenital Cardiovascular Anomalies that Cause Right Heart Failure

The cardiovascular anomalies that cause right heart decompensation are outlined in Table II. In the neonate, right heart failure is common. It is usually caused by persistent pulmonary hypertension of the newborn (persistent fetal circulation). The echocardiographic features of right heart failure in the newborn are, however, nonspecific. The right ventricle is dilated and hypertrophied (Fig. 3).

TABLE II

Primary Causes of Right Heart Failure (Due to Extracardiac or Cardiac Malformation)

- Extracardiac
  - Vein of Galen or other cerebral arteriovenous malformation
  - Hepatic arteriovenous malformation
- Cardiac
  - Ebstein's anomaly of the tricuspid valve
  - Total anomalous pulmonary venous connection
  - Uhl's anomaly
  - Postoperative congenital heart disease
    - Transposition of the great arteries after atrial switch (Mustard or Senning procedure)
    - Tetralogy of Fallot
    - Fontan for univentricular hearts
- Pulmonary hypertension secondary to congenital cardiac disease (Eisenmenger's complex)

Doppler estimation of right-side pressures indicates elevated right ventricular and pulmonary artery systolic pressures. A dilated superior vena cava, inferior vena cava, or coronary sinus may indicate an anomalous pathway of pulmonary venous return, site of a systemic arteriovenous malformation, severe tricuspid regurgitation, or right ventricular failure. A dilated superior vena cava (Fig. 4) should, however, alert the echocardiographer to total anomalous pulmonary venous return

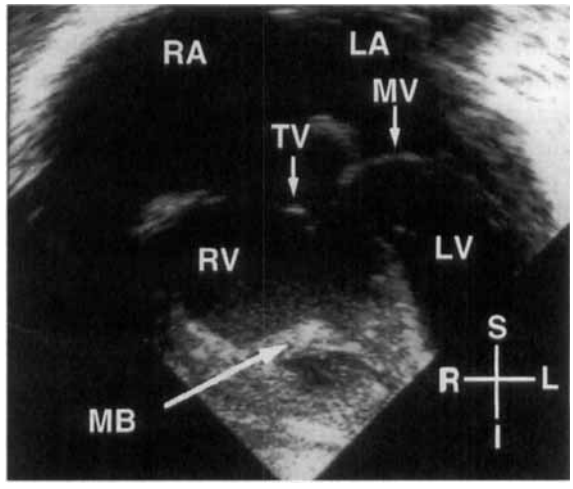

Figure 3. *Apical four-chamber cross section recorded from a newborn with right heart failure secondary to a hepatic arteriovenous malformation. The right ventricle (RV) is dilated and hypertrophied and compresses the left ventricle (LV). LA = left atrium; MB = moderate band; MV = mitral valve; RA = right atrium; TV = tricuspid valve. Compass: I = inferior; L = left; R = right; S = superior.*

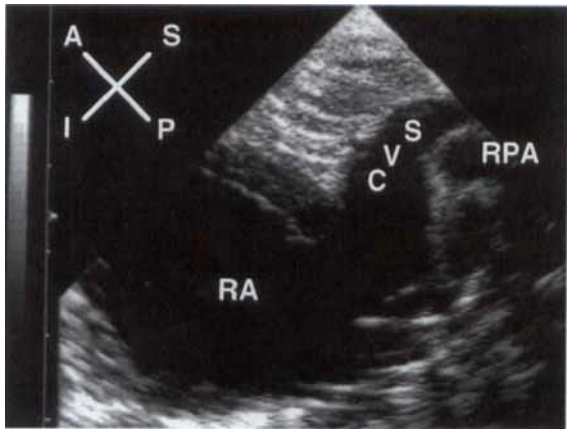

Figure 4. *Right parasternal image of a dilated superior vena cava (SVC) recorded from an infant with supracardiac total anomalous pulmonary venous connection. RA = right atrium; RPA = right pulmonary artery. Compass: A = anterior; I = inferior; P = posterior; S = superior.*

to the superior vena cava (Fig. 5) or a vein of Galen arteriovenous fistula (Fig. 6). The usual supracardiac pathway of anomalous pulmonary venous return is to a confluence of pulmonary veins behind the left atrium, a left vertical vein, the innominate vein, and, finally, the superior vena cava (Fig. 5). A sagittal or coronal scan of the brain through the anterior fontanelle will show a large echo-free space (venous varix) in the posterior fossa in infants with a vein of Galen fistula (Fig. 6A).[17] Color flow Doppler enables the ultrasonographer to outline the venous varix (Fig. 6B) or to trace the pathway of anomalous pulmonary venous return or vascular supply of the venous varix in the brain.

A dilated inferior vena cava (Fig. 7) may indicate total anomalous pulmonary venous connection below the diaphragm or a hepatic arteriovenous malformation (Fig. 8A). Anomalous pulmonary venous connection may drain to the portal vein, inferior vena cava, hepatic vein, or ductus venosus. Hepatic arteriovenous malformations may occur as a direct fistula, isolated hemangiomata, or diffuse hemangioendothelioma. Color Doppler provides the means for tracing the pathway of anomalous pulmonary venous drainage or for assessing the type of arteriovenous malformation (Fig. 8B).

A dilated coronary sinus is seen with total anomalous pulmonary venous return to the coronary sinus or a persistent left superior vena cava to the coronary sinus.

Ebstein's anomaly of the tricuspid valve may present as right heart failure in the newborn or older child (Fig. 9); however, there does not appear to be a relationship between echo-determined indices of right ventricular function and the New York Heart Association functional class in these patients.[18] In addition to morphological and functional abnormalities of the right ventricular myocardium, valvar abnormalities and arrhythmias contribute to right heart failure in these patients.

A rare congenital defect of the right ventricle is Uhl's anomaly, also called parchment right ventricle. These patients often live into adulthood and present with right heart failure secondary to inadequate right ventricular contraction. By cross-sectional imaging, a large right ventricle is observed along with paradoxical septal motion and an akinetic right ventricular free wall. In contrast to Ebstein's anomaly, the tricuspid valve leaflets are not displaced. Color Doppler will show varying degrees of tricuspid regurgitation.

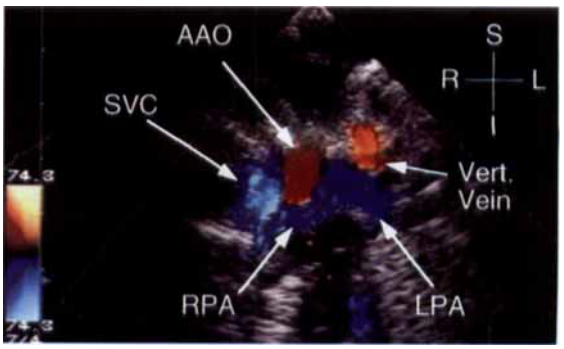

Figure 5. *Transverse suprasternal notch image displaying the pathway of venous return recorded from a newborn with supracardiac total anomalous pulmonary venous connection. From right (R) to left (L), flow is observed traveling in an inferior (I) direction in the superior vena cava (SVC), superior (S) direction in the ascending aorta (AAO), inferior direction in the left (LPA) and right (RPA) pulmonary artery, and superior direction in the vertical vein (Vert. Vein).*

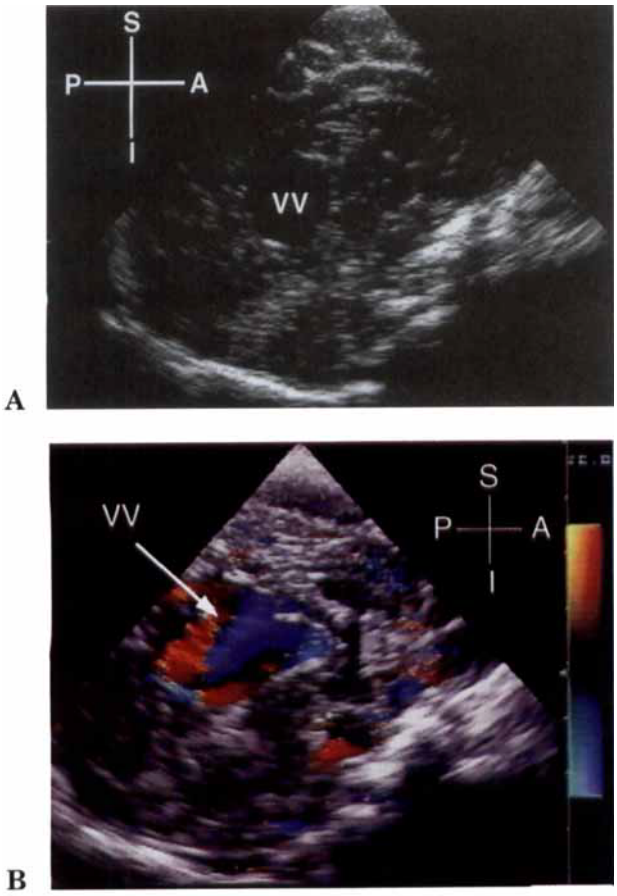

Figure 6. *(A) Sagittal view of the brain recorded through the anterior fontanelle from an infant with a vein of Galen fistula. VV = venous varix. Compass: A = anterior; I = inferior; P = posterior; S = superior. (B) Sagittal view of the brain recorded using color flow mapping. Within the venous verix (VV), swirling (red and blue) flow is observed. Color flow mapping identifies the echo-free space within the brain as vascular.*

Postoperative Congenital Heart Disease

It is not within the scope of this text to discuss all of the facets of postoperative congenital heart disease. However, it is common to observe right heart failure in three postoperative situations: transposition of the great arteries after atrial switch, tetralogy of Fallot, and the Fontan circulation.

Transposition of the Great Arteries

Redirection of pulmonary and systemic venous return at the atrial level (Mustard or Senning procedure) provides physiological correction of complete transposition of the great arteries. Thus, the morphological right ventricle remains the systemic arterial ventricle. Right ventricular shape, fiber orientation, and atrioventricular valve structure, however, may not be suitable for long-term survival. As these children grow into adulthood, progressive right ventricular failure and tricuspid valve regurgitation become problematic.[19,20] In these patients, two-dimensional (2-D) imaging has been shown to be useful in evaluating right ventricular ejection fraction,[21] and color Doppler provides important information regarding the degree of tricuspid regurgitation.

Tetralogy of Fallot

The majority of children with tetralogy of Fallot require a transannular patch across the right ventricular outflow tract as part of their repair. Thus, most children have some degree of pulmonary regurgitation and right ventricular volume overload after the repair. A small portion of these children not only have pulmonary regurgitation but also may have tricuspid regurgitation or residual outflow tract obstruction. In these patients, the combination of 2-D imaging in conjunction with color and continuous-wave Doppler al-

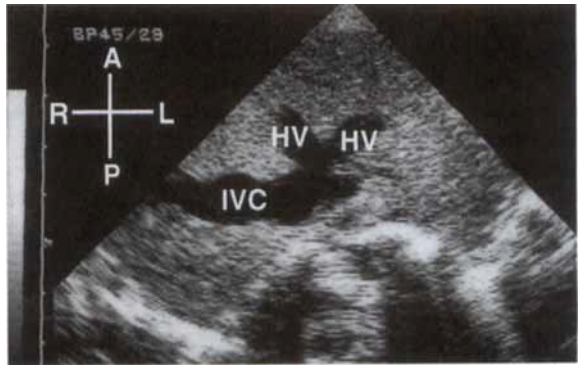

Figure 7. *Subxyphoid image of a dilated inferior vena cava (IVC) and hepatic veins (HV) recorded from an infant with a hepatic arteriovenous malformation. Compass: A = anterior; I = inferior; P = posterior; S = superior.*

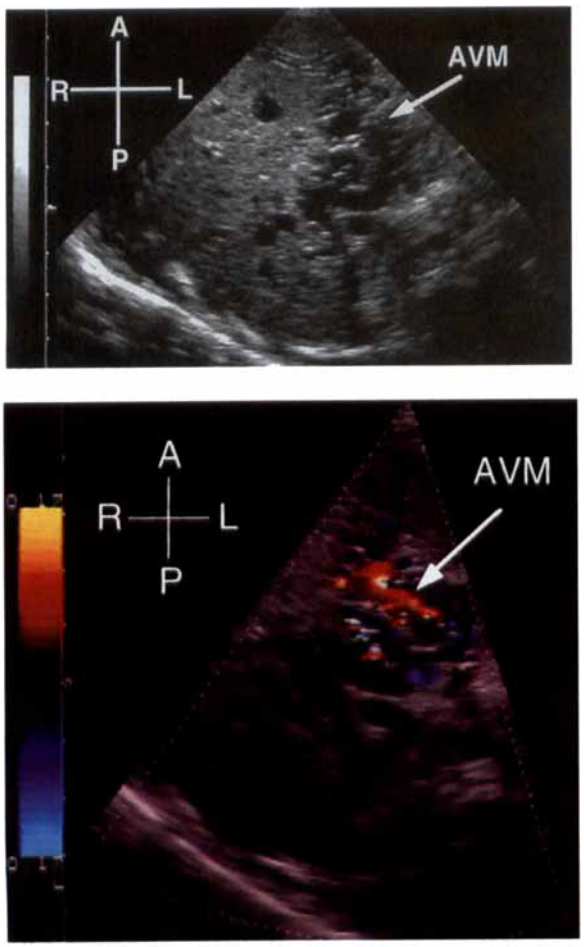

Figure 8. *(A) Transverse cross section of liver displaying the complex of vessels making up a hepatic arteriovenous malformation (AVM). Compass: A = anterior; I = inferior; P = posterior; S = superior. (B) Transverse cross section of liver with color flow mapping. The color map shows the characteristic turbulent flow observed in the AVM and identifies the echo-free space observed on Fig. 8A as a vascular complex.*

lows noninvasive assessment of the source of right heart failure.

The Fontan Circulation

Cavopulmonary right heart bypass is the procedure of choice in selected patients with univentricular heart. There are a variety of connections used by surgeons; the intra-atrial lateral tunnel and extracardiac conduit appear to be the most common surgical methods. Ob-

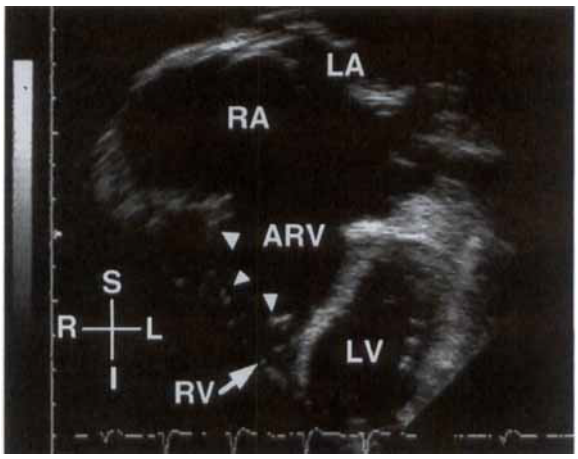

Figure 9. *Apical four-chamber image recorded from a patient with Epstein's anomaly. The typical displacement of the tricuspid valve (small arrows) creating the atrialized right ventricle (ARV) is seen. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle. Compass: A = anterior; I = inferior; P = posterior; S = superior.*

struction of flow through the Fontan pathway will lead to systemic venous hypertension and signs and symptoms of right heart failure. Transthoracic and transesophageal Doppler echocardiography[22] allows the imager to trace the pathway of the inferior vena cava to the pulmonary artery and of the superior vena cava to the pulmonary artery. Thus, the site of obstruction can be detected.

Secondary Right Heart Failure

We mentioned persistent fetal circulation as the most common cause of right heart failure in the newborn. This diagnosis is made after anatomic defects have been eliminated from consideration and there is Doppler evidence of pulmonary hypertension in conjunction with a right-to-left shunt at either the atrial or ductal level, or both.

Table III presents the differential diagnosis of right heart failure from pulmonary or associated airway diseases such as upper airway obstruction, thoracic cage abnormalities, neuromuscular disorders, and hypoventilation syndromes. The common denominator in these diseases is hypoxia with increased pulmonary

TABLE III

Secondary Causes of Right Heart Failure (Due to Airway Disease [Cor Pulmonale])

Parenchymal lung disease
Chronic obstructive lung disease (cystic fibrosis, asthma)
Restrictive lung disease (collagen vascular disease)
Combined obstruction/restrictive disease (bronchopulmonary dysplasia)
Persistent pulmonary hypertension of the newborn
Upper airway obstruction
Tonsil/adenoid hypertrophy
Craniofacial disorders, (e.g., Pierre-Robin anomalad)
Down's syndrome
Vascular ring
Throacic cage abnormalities (asphyxiating thoracic dystrophy)
Neuromuscular disorders (muscular dystrophies)
Respiratory control disorders
Pickwickian syndrome
Central hypoventilation
Sleep apnea syndrome
Pulmonary vascular diseases
Primary pulmonary hypertension
Pulmonary thromboembolism
Pulmonary veno-occlusive disease
Other unusual causes (high-altitude pulmonary hypertension)

Adapted from *Heart Disease in Infants, Children, and Adolescents*, 5th ed. Baltimore, Williams & Wilkins, chapter 101, pp. 1717-1724.

vascular resistance that leads to increased pulmonary artery pressures. As a consequence of the increased right ventricular afterload, there is progressive ventricular hypertrophy, dilation, and failure.

Noninvasive estimation of right heart failure is particularly useful in acute upper airway obstruction[23] when evaluating patients for failure due to pulmonary disease. The transesophageal approach should be considered when there is air trapping and hyperexpanded lungs that preclude the transthoracic approach.

Conclusion

Right heart failure in infants and children occurs as a consequence of a congenital defect or pulmonary or upper airway disease and as a sequela of surgical correction of congenital heart disease. A combined 2-D Doppler approach is useful in the assessment of right heart function and pulmonary hypertension.

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